Mucinous cystic neoplasm of the pancreas in a male patient

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Abstract

Mucinous cystic neoplasms (MCNs) make up a morphologic family of similar appearing tumors arising in the ovary and various extrapancreatic organs such as pancreas, hepatobiliary tract and mesentry. MCNs of the pancreas occur almost exclusively in women. Here, we report a rare case of MCN in a male patient. A 39-year-old man was admitted to our hospital with the chief complaint of back pain. Abdominal computed tomography revealed a multilocular cystic mass 6.3 cm in diameter in the pancreatic tail. In addition, the outer wall and septae with calcification were demonstrated in the cystic lesion. On magnetic resonance imaging, the cystic fluid had low intensity on T1-weighted imaging and high intensity on T2-weighted imaging. Endoscopic retrograde cholangio-pancreatography (ERCP) showed neither communication between the cystic lesion and the main pancreatic duct nor encasement of the main pancreatic duct. Endoscopic ultrasonography revealed neither solid component nor thickness of the septae in the cystic lesion. Consequently, we performed distal pancreatectomy with splenectomy under the diagnosis of cystic neoplasia of the pancreas. Histopathologically, the cystic lesion showed two distinct component: an inner epithelial layer and an outer densely cellular ovarian-type stromal layer. Based on these findings, the cystic lesion was diagnosed as MCN.

Introduction

Mucinous cystic neoplasms (MCNs) have been defined as large, separated, thick-walled cysts without connection to the pancreas duct system. Based on the WHO criteria in 1996, the study for 130 cases of MCN with ovarian-type stroma (OS) indicated to be female patients appearances in whole cases and bodytail location in almost. Therefore, as a rough rule for pancreas cystic neoplasms, in male and in the head of the pancreas are likely to be IPMNs, whereas cystic lesions in the bodytail in female may be either an MCN or IPMN. And, in the past several years, mucinous cystic neoplasms of the pancreas have been diagnosed more and more frequently. Then it has become crucial for physicians working in this field to have a clear understanding of the biology on these tumors. From recent our experience for a male case with MCN, which is quite rare, diagnostic evaluation and therapeutic procedures including surgical indication will be argued with the references.

Case Report

A 39-year-old man was admitted to our hospital with the chief complaint of back pain. There was no history of previous abdominal imaging examinations, and another abdominal episode, such as sudden abdominal pain, abdominal trauma, or abdominal operation. He was just social drinker of alcohol and was not smoker. On physical examination, abdomen was soft and flat, and no tenderness was noticed. Laboratory tests showed an elevation of AST, up to 39 IU/L (normal ¥ 35 IU/L), ALT, up to 56 IU/L (normal ¥ 40 IU/L). The other data including tumor markers (CEA, CA19-9 and DUPAN-2) were within normal range. Abdominal CT revealed a multilocular cystic mass, 6.3 cm in diameter, with wall calcification in the pancreatic tail (Figure 1). On MRI, the cystic lesion was hypointense in T1-weighted imaging and hyperintense in T2-weighted imaging with low intense capsule and septum (Figure 2a,b). ERCP showed neither communication between the cystic lesion and the main pancreatic duct nor encasement nor interruption of the main pancreatic duct. Endoscopic ultrasonography revealed neither solid component nor thickness of the septae in the cystic lesion. Consequently, under the diagnosis of neoplastic cyst of the pancreas, such as lymph epithelial cyst, serous cystic neoplasm, branched-type lesion of IPMN, cystic change of endocrine tumor, and epidermoid cyst derived from accessory spleen in the pancreas, distal pancreas with spleen was removed. Macroscopically (Figure 3), the cystic lesion, measuring for 6.5x5.8x5.2 cm in size, was round with a smooth surface and was surrounded by normal pancreatic tissue. The cut surface demonstrated a multilocular cystic pattern containing thick yellowish mucin, and the lesion was surrounded by a fibrous capsule.

Figure 1. Abdominal CT revealed a multilocular cystic mass, 6.3 cm in diameter, with wall calcification and no solid component in the pancreatic tail.

Figure 2. Magnetic resonance imaging showed that the cystic lesion was hypointense in T1-weighted imaging (a) and hyperintense in T2-weighted imaging with low intense capsule and septum (b).
The cystic lesion showed two distinct components; an inner mucinous epithelial layer and an outer densely cellular stromal layer. The mucinous epithelium showed no cytological atypia in the epithelial layer and no infiltrate into the stromal layer. This stromal layer was consisted of spindle-shaped cells with round to oval nucrei and a small amount of cytoplasm, suggesting the finding for OS.

Microscopically (Figure 4 a,b), the cystic lesion showed two distinct component; an inner mucinous epithelial layer and an outer densely cellular stromal layer. The mucinous epithelium showed no cytological atypia and did not infiltrate into the stromal layer. This stromal layer was consisted of spindle-shaped cells with round to oval nucrei and a small amount of cytoplasm, suggesting the finding for OS.

Discussion

Mucin-producing cystic neoplasms of the pancreas have developed a well-recognized entity. In the two decades, due to the utility of high-resolution abdominal imaging techniques, similar cystic lesions of the pancreas are increasingly identified incidentally, and a large number of patients have undergone surgical resection. In 1996, under the aim to describe and categorize the cystic lesions of the pancreas, the WHO classification defined MCN as cystic epithelial neoplasms composed of columnar, mucin-producing epithelium, supported by OS. The OS is known for forming a band of densely packed spindle cells beneath the epithelium; its presence has become a critical requirement as MCN. Then, the MCN was estimated for the different concept from IPMN in the past categories for cystic lesion. And, the Armed Forces Institute of Pathology (AFIP) classification also added the finding for no communication with the pancreas ductal system. Taken together, no doubt to diagnose the present cystic lesion as MCN was detected. Although the developmental process of MCN has not been well understood, it is indicated to originate from remnant primordial gonadal cells that migrated to the pancreas, because the left primordial gonad and dorsal pancreas anlage lie side by side during embryogenesis. The dorsal anlage develops the body and tail of the pancreas; therefore the MCN was frequently raised in the dorsal pancreas and detected in female. Indeed, from the past reports for 130 cases, and 57.7% in invasive carcinoma cases, and 100% in adenoma to minimally invasive carcinoma, the 5-year survival rate of MCN patients was estimated for the different concept from IPMN in the past categories for cystic lesion. And, the Armed Forces Institute of Pathology (AFIP) classification also added the finding for no communication with the pancreas ductal system. Taken together, no doubt to diagnose the present cystic lesion as MCN was detected. According to the recent research for pancreatic cystic tumors by the Japan Pancreas Society (1992-2001), detected 179 cases with MCN were female in whole. To diagnose the cystic lesion for MCN, the presence of OS pattern should be important; if limiting the tumor with OS but not without OS, all male cases were excluded. Therefore, it is likely that many of the MCN cases reported in male in the early literature were IPMN or other cystic lesion. To the best of our knowledge, 2 male cases with WHO cri-

References


